



Submitted: 22.08.2024
Accepted: 14.10.2024
Early publication date: 09.12.2024

Endokrynologia Polska
DOI: 10.5603/ep.102204
ISSN 0423–104X, e-ISSN 2299–8306

Riedel's thyroiditis requiring an emergency surgery: rare manifestation of IgG4-related disease

Przemysław Paul¹, Andrzej Hellmann², Jolanta Szade³, Joanna Rejszel-Baranowska⁴, Łukasz Obolończyk¹

¹Department of Occupational, Metabolic, and Internal Diseases, Medical University of Gdańsk, Gdynia, Poland

²Department of General, Endocrine, and Transplant Surgery, Medical University of Gdańsk, Gdańsk, Poland

³Department of Pathomorphology, Medical University of Gdańsk, Gdańsk, Poland

⁴Department of Cardiology and Internal Medicine, Medical University of Gdańsk, Gdynia, Poland

Key words: Riedel's thyroiditis, Riedel's goitre, IgG4-related disease

Riedel's thyroiditis (RT) is a rare fibrotic process of unknown aetiology that involves the thyroid and often extends beyond the gland into the surrounding tissues. Nowadays, RT is considered a part of the IgG4-related disease (IgG4-RD) spectrum [1]. Due to the rarity of the condition and lack of randomised controlled trials, there are no diagnostic or therapeutic algorithms concerning management of RT [2].

In this article we present the case of a patient with RT, who required an emergency surgery. A 59-year-old male with a past medical history of heart failure with reduced ejection fraction, atrial fibrillation, chronic kidney disease, type 2 diabetes, chronic obstructive pulmonary disease, and hypothyroidism (on supplementation of levothyroxine 175 µg per day), attended the emergency department with exertional dyspnoea, shortness of breath, wheezing, and hoarseness. The symptoms had been ongoing for several months with significant deterioration within the 2 days prior to the admission. Physical examination showed a "stony" hard, non-tender, painless goitre with asymmetrically enlarged left lobe of the thyroid, stridor, and features of superior vena cava syndrome such as oedema of the face and positive Pemberton's sign. The laboratory tests exposed subclinical hypothyroidism with a thyroid-stimulation hormone (TSH): 5.739 mIU/L (reference range [RR], 0.42–3.8 mIU/L), mildly elevated C-reactive protein (CRP): 28.6 mg/L (RR, < 5.0 mg/L), and complete blood count was within RR. Computed tomography (CT) revealed an enlarged thyroid gland with numerous calcifications within the thyroid tissue. Its left lobe reached the upper mediastinum. The trachea was compressed by the goitre to 3 mm in the coronal dimension in

the transverse plane at the narrowest point. No lymphadenopathy was found. Anaplastic thyroid cancer was suspected. The patient was qualified for an emergency surgery. During operation the thyroid tumour was assessed unresectable. The procedure was limited to the exposure of malacic trachea and partial removal of the right lobe of the thyroid. A tracheostomy tube was inserted. Pathological examination of the obtained material revealed no neoplasia. The thyroid tissue was almost completely overtaken by fibrosis and hyalinosis. The process extended beyond the thyroid and affected muscles, adipose tissue, and the trachea. A large number of IgG4-positive plasma cells were found in the histologic sections (> 10 IgG4+/high-power field). The patient was later admitted to the internal medicine department for further disease evaluation and treatment. The serum IgG4 concentration was within RR. Noteworthy, elevated serum IgG4 levels are observed in approximately 75% of IgG4-RD cases and are not necessary for the diagnosis [3]. Hypoparathyroidism, which can accompany RT, was excluded [3]. The patient had elevated both anti-thyroid peroxidase antibodies (anti-TPO) and anti-thyroglobulin antibodies (anti-TG), which are observed in 43% and 27% of patients with RT, respectively [2]. The CT of the head, thorax, and abdomen revealed no features of retroperitoneal or orbital fibrosis. In the apex of the left lung, an enhancing nodular lesion with soft tissue density and the size of 26 × 23 × 20 mm was found. The morphology and dimensions of this finding were compared with the previous, available CT examinations. The tumour was stable for at least 4 years. This suggests that the lesion might be a part of the IgG4-related respiratory disease [4]. More-



Przemysław Paul, MD, Medical University of Gdańsk, Department of Occupational, Metabolic, and Internal Diseases, ul. Powstania Styczniowego 9b, 81–519 Gdynia, Poland, phone: +48 58 699 84 02, e-mail: ppaul@gumed.edu.pl



Figure 1. Imaging studies. **A.** Chest X-ray showing the compressed trachea (arrow); **B.** Ultrasound scan of the left lobe of thyroid gland showing massive calcifications (arrow) within thyroid tissue; **C.** Computed tomography of the neck and chest showing goitre with massive calcifications within thyroid tissue (black arrow) and compressed trachea (white arrow); **D.** Computed tomography of the neck and chest showing the lung tumour as a manifestation of the IgG4-related respiratory disease

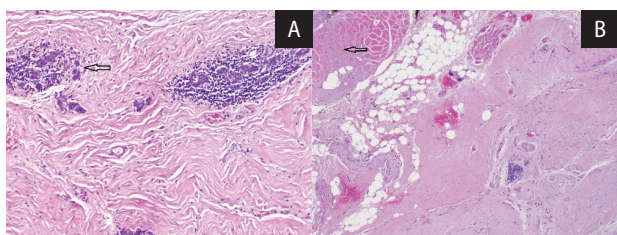


Figure 2. Microscopic pictures. **A.** Destruction and atrophy of the thyroid follicles (arrow), focal inflammation, severe fibrosis [haematoxylin and eosin (HE) staining, magnification 100 \times]; **B.** Dense fibrous tissue involving thyroid gland and extending in the adjacent skeletal muscle (arrow) (HE staining, magnification 40 \times)

over, enlargement of the thyroid gland and narrowing of the trachea was also described in the previous CT, but due to lack of symptoms the patient had resigned from further diagnostics. Glucocorticoids (GCS) are considered the first-line therapy of IgG4-RD [5]. Due to the multimorbidity of the patient, methylprednisolone pulse therapy was started. A total dose of 3000 mg was administered within 4 weeks. After the sixth pulse of GCS out of 12 planned, no remarkable response was achieved. Furthermore, the patient developed severe, bilateral pneumonia with pleural empyema of the right pleural cavity. In consequence, he required pleural drainage and several weeks of antibiotic treatment. Due to the advanced stage of the disease, multimorbidity, and serious complications of the GCS therapy, no alternative immunosuppressive treatment was started. The patient has been scheduled for regular follow-up visits in the ambulatory care, where thyroid function has been regularly assessed.

Our clinical case illustrates the difficulties in the management of RT and IgG4-RD. Complex assess-

ment of the patient and observation for other manifestations of IgG4-RD is required. The cooperation of multiple specialists including endocrinologists, surgeons, radiologists, and pathologists seems to be essential. Apart from GCS, other agents such as mycophenolate mofetil, rituximab, or tamoxifen can be used to suppress RT [3]. Our clinical case shows that the decision about starting immunosuppressive therapy has to be made carefully, especially when the disease seems to be very advanced.

Authors' contributions

Conception of the article: PP and L.O. Design of the article: PP, A.H., J.S., J.R.B., and L.O. Acquisition of data: PP, A.H., J.S., J.R.B., and L.O. Analysis and interpretation of data: PP, A.H., J.S., J.R.B., and L.O. Writing of the manuscript: PP, A.H., and J.S. Revising of the manuscript: PP and L.O.

Acknowledgments

We would like to thank our patient in this clinical vignette.

Conflict of interest

The authors have no conflicts of interest to declare.

References

- Dahlgren M, Khosroshahi A, Nielsen GP, et al. Riedel's thyroiditis and multifocal fibrosclerosis are part of the IgG4-related systemic disease spectrum. *Arthritis Care Res (Hoboken)*. 2010; 62(9): 1312–1318, doi: [10.1002/acr.20215](https://doi.org/10.1002/acr.20215), indexed in Pubmed: [20506114](https://pubmed.ncbi.nlm.nih.gov/20506114/).
- Zala A, Berhane T, Juhlin CC, et al. Riedel Thyroiditis. *J Clin Endocrinol Metab*. 2020; 105(9), doi: [10.1210/clinem/dgaa468](https://doi.org/10.1210/clinem/dgaa468), indexed in Pubmed: [32687163](https://pubmed.ncbi.nlm.nih.gov/32687163/).
- Czarnywojtek A, Pietrończyk K, Thompson LDR, et al. IgG4-related sclerosing thyroiditis (Riedel-Struma): a review of clinicopathological features and management. *Virchows Arch*. 2023; 483(2): 133–144, doi: [10.1007/s00428-023-03561-2](https://doi.org/10.1007/s00428-023-03561-2), indexed in Pubmed: [37204493](https://pubmed.ncbi.nlm.nih.gov/37204493/).
- Zen Y, Inoue D, Kitao A, et al. IgG4-related lung and pleural disease: a clinicopathologic study of 21 cases. *Am J Surg Pathol*. 2009; 33(12): 1886–1893, doi: [10.1097/PAS.0b013e3181bd535b](https://doi.org/10.1097/PAS.0b013e3181bd535b), indexed in Pubmed: [19898222](https://pubmed.ncbi.nlm.nih.gov/19898222/).
- Zhang W, Stone JH. Management of IgG4-related disease. *Lancet Rheumatol*. 2019; 1(1): e55–e65, doi: [10.1016/S2665-9913\(19\)30017-7](https://doi.org/10.1016/S2665-9913(19)30017-7), indexed in Pubmed: [38229361](https://pubmed.ncbi.nlm.nih.gov/38229361/).